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Echocardiographic findings in newborns and postneonatal infants undergoing preoperative evaluation for surgically correctable non-cardiac congenital malformations

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Abstract*: Background:* Congenital cardiac malformations could co-exist with surgically correctable non-cardiac congenital structural abnormalities. The occurrence of the two conditions portends increased anaesthetic risk and perioperative complications. Early recognition could favourably alter the post-operative outcome.
Objectives: To describe the prevalence and pattern of congenital heart defects in young infants with extracardiac structural malformations.
Methods: Consecutive newborns and postneonatal infants with congenital malformations being prepared for surgery were sent to the paediatric cardiology unit of University of Abuja Teaching Hospital for echocardiography and were recruited over a 30moth period. A transthoracic echocardiography, following the guidelines of the American Society of Echocardiography, was used to evaluate the cardiac structures.
Results: Of the 44 newborns and postneonatal infants with congenital non-cardiac malformations, 27 (61.4%) were males while 17 (38.6%) were females, giving a M:F ratio of 1.59:1. Gastrointestinal malformations constituted 47.7% of cases. Omphalocoele major and biliary atresias were the most common malformations seen. Seven babies had completely normal heart, five had isolated patent foramen ovale, thus giving a total of 12 babies with structurally normal hearts. Of the 32 babies with structural heart defects, 17(53.1%) had single defect while 15 (46.9%) had two or more defects consisting of 26 acyanotic and 4 cyanotic defects. A total of 47 cardiac defects were seen. Forty-two (89.4%) of the heart defects were simple while 5(10.6%) were complex. Atrial septal defect 13,(27.7%), patent ductus arteriosus 14, (29.8%) and ventricular septal defect 7,(14.9%) were the commonest simple heart defects while atrioventricular septal defects predominate among the complex category
Conclusions: Congenital structural abnormalities are common indications for surgical interventions in young infants. Recognition of coexisting structural cardiac abnormalities is essential in preoperative planning and may be a useful guide in averting untoward anaesthetic accident.

Key words: Newborns, infants, congenital heart defects, extracardiac malformations

Introduction

Extracardiac structural malformations account for significant part of surgical consultations in the neonatal and immediate postneonatal period mainly due to the grotesque appearance of those with the malformations especially omphalocoele major, myelomemningocoele and bladder extrophy, as well as the unstable presentation of newborns with major internal malformations such as tracheosophageal fistula, diaphragmatic hernia and intestinal atresias. Non-cardiac malformation was reported to be the immediate or major cause of death in 12% of newborns in a study by Jaiyemisi and Antia.[1] The contribution of accompanying cardiac malformations to the mortality is however unknown.

Neonates and infants with congenital heart defects have a two-fold increase in mortality from non-cardiac sur-

gery.[2] Anaesthesia-related paediatric cardiac arrest, according to a review, occurred in 75% of patients under 2 years of age with cardiac defects during non-cardiac surgery.[2,3] Nevertheless, in experienced hands, anaesthesia for neonates with complex heart diseases presenting for treatment of general surgical emergencies can still be conducted with manageable complications.

Surgical amendment of the seemingly frightening external congenital malformation has greatly altered the outcome of management and the acceptance of the baby by parents. However, the management and prognosis of the patient may be determined by the presence or otherwise of cardiac malformations especially at the perioperative period where the cardiac defect may constitute a significant anaesthetic risk and delay postoperative recovery.
The present study aimed at describing the prevalence and pattern of congenital heart defects in young infants with non-cardiac malformations preoperatively.

Methodology

Consecutive newborns and postneonatal infants being prepared for surgery for non-cardiac malformations were sent for echocardiographic evaluation and were recruited for the study between March, 2015 and September, 2017.

Echocardiography was performed using a portable General Electric Vivid e echo machine fitted with a 6S (5-7MHz) transducer. Standard two-Dimensional, M-mode, and Doppler echocardiograms were performed in the supine and left lateral decubitus positions and measurements done as recommended by the American Society of Echocardiography.
Ethical clearance was obtained for a study on structural heart diseases in children and this study is part of a larger one.

Results

A total of 44 newborns and postneonatal infants with non-cardiac malformations were referred for echocardiography; 27 (61.4%) were males while 17 (38.6%) were females, giving a male-to-female ratio of 1.59:1. Eighteen (40.9%) of the patients presented within the first week of life. Details of the age at presentation are displayed in Table 1 which also shows the gender distribution of the patients studied.

Table 1: Age and sex distribution of 44 young infants with extra-cardiac malformations

Age	Gender		Total
	Male (n = 27) n (%)	Female (n = 17) n (%)	N (%)
0-7days	11 (40.7)	7 (41.2)	18 (40.9)
8-14 days	4 (14.8)	3 (17.6)	7 (15.9)
15-28 days	4 (14.8)	3 (17.6)	7 (15.9)
>28 days	8 (29.6)	4 (23.5)	12 (27.3)

Abnormalities of the digestive system constituted 47.7% of the extracardiac malformations seen in the patients. Omphalocoele major and intestinal atresias were the most common of this malformation followed by biliary atresia. Syndromic malformations and respiratory system anomaly contributed 18.2 and 11.4% respectively. Pattern of congenital malformations in the patients is shown on Table 2.

Table 2: Pattern of extracardiac malformations among patients studied

Type of extracardiac defects	Frequency	Percentages
Digestive System	21	47.7
Omphalocoele	7	15.9
Intestinal atresias + imperforate anus	7	15.9
Biliary atresia	4	9.0
Inguinal hernia	1	2.3
Cleft lip and palate	2	4.5
Syndromic Malformation	8	18.2
Down Syndrome	4	9.1
Pierre Robin Syndrome	2	4.5
Edward Syndrome	1	2.3
Turner's Syndrome	1	2.3
Respiratory System	5	11.4
Tracheosophageal fistula	3	2.9
Diaphragmatic hernia	1	2.9
Laryngomalacia	1	2.9
Genitourinary System	3	6.8
Bladder estrophy	2	4.5
Posterior urethral valve	1	2.3
Central nervous system	2	4.5
Thoracic myelomeningocoele	1	2.3
Lumbosacral meningocoele	1	2.3
Skeletal System	4	9.1
Polydactyly	1	2.3
Multiple limb malformations	2	4.5
Achondroplasia	1	2.3
Vascular	1	2.3
Giant haemangioma	1	2.3
Total	44	100

The prevalence of cardiac defects among infants with gastrointestinal anomaly was statistically significant, (χ^2= 6.380, p = 0.029). However the association of cardiac defects with other non-cardiac malformations failed to reach statistical significance, Table 3.
Seven babies had completely normal heart, 5 had isolated patent foramen ovale, thus giving a total of 12 babies with structurally normal hearts. Of the 32(72.7%, 32/44) babies with structural heart defects, 17 (53.1%) had single defect while 15 (46.9%) had two or more defects consisting of 26acyanotic and 4 cyanotic defects. A total of 47 cardiac defects were seen. Forty-two (89.4%) of the heart defects were simple while 5 (10.6%) were complex heart defects. Atrial septal defect (13, 27.7%), patent ductus arteriosus (14, 29.8%) and ventricular septal defect (7, 14.9%) were the commonest simple heart defects while the comple category consists of atrioventricular septal defect, double outlet right ventricle, truncus arteriosus, mitral atresia with single ventricle anatomy and total anomalous pulmonary venous return.

Distribution of cardiac defects is shown on Table 4 and 5.

Table 3: Distribution of extra-cardiac anomaly among infants with congenital heart diseases

Extracardiac anomaly	Congenital heart disease		Total	χ^2	P
	Present *n=32*	Absent *n=12*			
Digestive system	19 (59.4)	2 (16.7)	21	6.380	0.029
Identified syndrome	7 (21.9)	3 (25.0)	10	0.000	1.000
Respiratory system	2 (6.25)	3 (25.0)	5	1.469	0.225
Genitourinary system	2 (6.25)	1 (8.33)	3	FET	1.000
Central nervous system	-	2 (16.7)	2	FET	0.069
Others (Skeletal, haematologic)	2 (6.25)	1 (8.33)	3	FET	1.000

FET = Fishers exact test

Table 4: Echocardiographic diagnosis in 44 infants with extra-cardiac malformations

Echocardiographic diagnosis	System involved in extracardiac malformations					
	Digestive	G.U.S	Syndromic	Respiratory	CNS	Others$\delta\delta$
Atrial Septal Defects	6	1	2	1	-	3
Ventricular Septal defects	5	-	2	-	-	-
Patent Ductus Arteriosus	4	-	6	1	-	3
Pulmonary Stenosis	2	1	-	-	-	-
Patent Foramen Ovale	3	1	1	-	-	-
Normal	2	1	3	3	2	1
Others	5**	-	$3^{\beta\beta}$	-	-	-

****** refers to Truncus arteriosus (1), Double outlet right ventricle (1), Total anomalous pulmonary venous drainage (1), Mitral atresia (1), Aortic stenosis (1)
ββ refers to Atrioventricular septal defect (2), Left superior vena cava (1)
δδ included giant haemangioma, skeletal abnormalities
G.U.S = Genitourinary system, CNS = Central nervous system

Table 5: Types of non-cardiac malfor mations seen in patients with complex cardiac defects

Non-cardiac malformation	Complex heart lesion
Down syndrome	Atrioventricular septal defect
Turner syndrome	Atrioventricular septal defect
Omphalocoele major	Double outlet right ventricle
Omphalocoele major	Mitral atresia with single ventricle anatomy
Diaphragmatic hernia	Truncus arteriosus

Discussion

Anomaly of the digestive system constituted the majority of abnormalities seen in the present study, similar to the observation made by Oyati*et al*[4]in Zaria but differs from that of Jaiyesimi and Antia[1] in Ibadan in which the prevalence of digestive system anomalies (23%) was superseded by musculoskeletal malformations (27%). On the overall, anomaly of the digestive system such as omphalocoele major were common in this subset of infants and tended to presents earlier, probably due to the grotesque appearance of the baby.[5-7]

The prevalence of cardiac defects among infants with extra-cardiac congenital malformations in the present study is high (72.7%), twice the prevalence recorded in a similar cohort in Zaria. The higher prevalence may be due to differences in study population. The Zaria study included children up to the age of 11years and it was retrospective. In the series of Gonzalez *et al*[5] from California, 50% of neonates with congenital heart defects also had extracardiac anomaly, thus corroborating the finding in the present study.

The prevalence of cardiac anomaly in neonates with external non-cardiac malformations in this study supports the need for preoperative echocardiography for this category of infants. Preoperative echocardiography helps to identify the presence and type of cardiac defects as well as the severity of the malformations and hence assists the surgeon, anaesthesiologist and neonatal cardiologist in weighing the risk-versus-benefit of the intended surgery, vis-à-vis the ability of the patient to withstand the stress of surgery and even more critical, the stress of anaesthesia.

Simple shunt defects were the most common type of cardiac defects in children with extra-cardiac malforma-

tions in this study. Atrial septal defects, ventricular septal defects and patent ductus arteriosus predominated among the simple heart lesions seen. Similar findings have been reported elsewhere.[1,4-7] There is however no established association between type of extra-cardiac anomaly and specific simple cardiac defect.

Complex cardiac defects were relatively rare, 10.6% of all heart defects seen, and occurred in infants with syndromic malformations and major gastrointestinal anomaly. Although relatively rare, the presence of complex cardiac defects in newborns with major gastrointestinal anomaly portends a precarious situation as the duo spells unfavourable outcome if not properly managed. However early identification of complex cardiac defects helps in planning for the best treatment option within the ambit of available resources.

The spectrum of cardiac defects documented in this study provides a guide in prognosticating the perioperative outcome of newborns and postneonatal infants undergoing surgery for non-cardiac malformations.

Conclusions

Cardiac malformations are common in young infants presenting with extra-cardiac surgical disorders. All newborns and immediate postneonatal infants undergoing corrective or palliative surgery for non-cardiac malformations should have cardiac evaluation before commencement of the surgery.

Conflict of interest: None
Funding: None

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